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Use of epidural anaesthesia for surgery in a patient with Kennedy's disease

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Use of neuraxial block in a patient with motor neuron disease is controversial. We describe the anaesthetic management by epidural anaesthesia of a patient with Kennedy's disease, a rare lower motor neuron disease characterized by progressive weakness and wasting of limbs and bulbar muscles. The perioperative course was uneventful, and there was no exacerbation of neurologic signs or symptoms. We suggest that a patient with Kennedy's disease may be successfully managed by epidural anaesthesia for surgical internal urethrotomy.

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Kennedy's disease is an X-linked lower motor neuron disorder characterized by progressive weakness and wasting of limbs and bulbar muscles.[1] We describe the management by epidural anaesthesia of a patient with Kennedy's disease. Searching Medline data from 1966 to May 2003 revealed no previous case reports of anaesthetic management of this disease.

Case report

A 57-yr-old, 61 kg, 163 cm, man was undergoing an internal urethrotomy for urethral stenosis. He had first noticed muscle weakness in the proximal part of the lower limbs at age 50. Six years later, he was diagnosed as suffering from Kennedy's disease. Physical examination revealed atrophy, weakness and fasciculations in the lower and upper limb muscles, and diminished deep tendon reflexes in the lower and upper extremities. He had slurred speech and slight difficulties in swallowing. Other findings included fasciculation and atrophy of the tongue and facial muscles. Gynecomastia was also apparent. Needle electromyogram (EMG) revealed large polyphasic motor units in the affected muscles, which were consistent with the diagnosis. Laboratory studies included a plasma creatine kinase concentration of 1223 u litre^{-1} (normal range 43–120 u litre^{-1}).

After detailed discussion and with the agreement of the patient, management by epidural anaesthesia was chosen. The patient was premedicated with roxatidine (H_2-blocker), 75 mg, post-orally. An epidural catheter was inserted at the L3–4 interspace and directed 5 cm cephalad. Epidural anaesthesia was established by injecting 10 ml of mepivacaine 2% via the catheter after a test dose of 3 ml of 2% mepivacaine, achieving a sensory block extending to T10. Oxygen was administered through a facemask at 5 litre min^{-1}. An additional increment of mepivacaine 2% (5 ml) was given epidurally during the operation. Throughout this period, ECG was normal, Sp_{O_2} greater than 98%, and the patient had no respiratory discomfort. The postoperative course was uneventful, and there was no exacerbation of neurologic signs or symptoms.

Discussion

Kennedy's disease is a recessive X-linked adult-onset form of motor neuron disease that is linked to a CAG repeat enlargement within the first exon of the androgen receptor gene.[2 3] The age of onset of Kennedy's disease is in adolescence and associated with symptoms such as gynecomastia, muscle pain, and premature muscular exhaustion. The number of CAG repeats appears to be correlated with the age of onset of weakness but not with the age of onset of Kennedy's disease.[4] There is a great variability in phenotypical expression and heterogeneity in clinical presentation since the severity of Kennedy's disease is not related to the size of the mutation.[4] Therefore, the

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diagnosis of Kennedy's disease may be missed and, as a consequence, its prevalence underreported.[5] The two findings distinguishing this disorder from amyotrophic lateral sclerosis are the absence of signs of pyramidal tract involvement and the presence of a subtle sensory neuropathy. Identification of a mutation of the androgen receptor gene aids in the diagnosis of Kennedy's disease, especially in mildly affected and sporadic cases.[2]

The problem areas associated with anaesthetic management of patients with Kennedy's disease are not fully clarified. However, patients with bulbar involvement with dysfunction of pharyngeal muscles may be predisposed to regurgitation and aspiration. General anaesthesia depresses the swallowing reflex and may further increase the risk of pulmonary aspiration.[6] The use of neuromuscular blocking agents is also a major concern in anaesthetic management in a patient with motor neuron disease. Although, in Kennedy's disease, it is not clear whether succinylcholine causes a hyperkalaemic response, it seems that the use of succinylcholine should be avoided because of possible unpredictable side effects. Decreased levels of acetylcholine may increase a patient's sensitivity to non-depolarizing neuromuscular blocking agents in motor neuron disease.[7] Taking into account the various disadvantages of general anaesthesia and the use of neuromuscular blocking agents in patients with motor neuron diseases, we chose epidural anaesthesia for our patient.

When neuraxial block is chosen, it is important to select an appropriate level as a sensory block above T6 decreases the expiratory reserve volume, leading to reduced clearance of secretions from the airway.[8] We used 3 ml of mepivacaine 2% as a test dose and then followed by 10 ml. The highest anaesthetic level in our case was T10 and the patient experienced no respiratory discomfort.

Exacerbation of pre-operative neurological impairment after neuraxial block has been reported.[9] However, there have been no reports of deterioration of pre-existing neurological function after epidural anaesthesia in patients with lower motor neuron diseases. Epidural anaesthesia has been successfully administered to patients with amyotrophic lateral sclerosis without neurological exacerbation or impairment of pulmonary function.[10]

In summary, we managed a patient with Kennedy's disease by epidural anaesthesia without exacerbation of neurological signs or symptoms. Although there is no evidence that any specific anaesthetic technique or drug in particular is best for patients with this disease, we suggest that it may be successfully managed with epidural anaesthesia in the surgical procedure of an internal urethrotomy.

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